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Case Report: Congenital Retroperitoneal Fibrosarcoma

Caroline M. Hamm, MD, Allan Pyesmany, MD, FRCP(C), and Lothar Resch, MD, FRCP(C)

We report a case of congenital retroperitoneal fibrosarcoma presenting in a 17-day-old male in which surgical tumor debulking was followed by chemotherapy. His clinical course was complicated. Intraoperatively, 80% of the tumor was removed and he experienced massive blood loss and significant cardiac arrhythmias. Following surgery he received chemotherapy and suffered life-threatening hepatic, hematological and gastrointestinal toxicity. The treatment resulted in clinical remission after 9 months of follow-up. A review of the current literature revealed the changing opinion of the optimal treatment of fibrosarcomas in children. Chemotherapy is playing an increasingly important role in the treatment of fibrosarcoma, both preoperatively and postoperatively.

Key words: chemotherapy, fibrosarcoma, retroperitoneal

INTRODUCTION

Fibrosarcoma in infants and children carries a substantially better prognosis than in adults [1,2]. It most frequently arises in the distal extremities with retroperitoneal disease being relatively unusual. A case of congenital retroperitoneal fibrosarcoma in a 17-day-old male is presented, in which surgical tumor debulking was followed by chemotherapy, resulting in clinical remission after 9 months of follow-up.

A review of the current literature reflects the changing opinion of the optimal treatment of fibrosarcomas in children. Chemotherapy is playing an increasingly important role in the treatment of fibrosarcoma today, despite previous opinion that total surgical excision offered the only effective treatment for sarcomas. Preoperative chemotherapy to reduce tumor size may prevent mutilating surgery [3]. Success with postoperative chemotherapy has recently been reported [3,4] as has chemotherapy alone [3,5–8].

CASE PRESENTATION

A 17-day-old male infant was admitted to the IWK Children's Hospital with the sole complaint for 1 week of projectile vomiting occurring 10 minutes following feedings. He had no other symptoms and was afrebrile. Bowel movements were normal. There had been no other vomiting except postfeeds. The patient was the product of an uneventful pregnancy and delivery and weighed 4.6 kilograms at birth. Circumcision at age 7 days resulted in local skin infection which responded to Amoxil. Antenatal ultrasonography had been normal on two occasions. The patient has a healthy 2-year-old brother. One paternal grandmother has carcinoma of the breast. There is no other family history of malignancy.

The pertinent physical finding was a large abdominal mass which extended from above the umbilicus on the right side, into the pelvis and an ill-defined area of firmness on the upper medial aspect of the right thigh. Laboratory investigations revealed a white blood cell count of 21.2×10^9 cells per litre, hemoglobin 144 gram per litre, platelets 29×10^9 cells per litre and no peripheral blasts. Bone marrow biopsy and aspirate were normal.

Ultrasonography disclosed a 10 by 8 centimeter solid, noncalcified pelvic mass. No calcifications were noted in the mass. A spastic but functional pylorus and bilateral hydronephrosis were noted. Both testes were normal. Computed tomography of the patient's abdomen (Fig. 1) substantiated the results of the ultrasound examination. The mass was partially necrotic, was not attached to the kidneys and appeared to be attached posteriorly to the mesenteric root. A bone scan showed increased uptake in the tumor region only. A gallium-67 scan identified a solid and necrotic tumor arising from the pelvis and no other areas of uptake. Chest x-ray, cerebrospinal fluid examination and computed tomography of the chest were normal. Multiple percutaneous needle biopsies of the tumor were taken during the same operation as insertion of the portacath. The biopsies offered the diagnosis of fibrosarcoma.

At age 24 days, the patient underwent subtotal resection of the mass. Surgery was complicated by excessive blood loss and several episodes of hypotension and car-

From the Departments of Hematology/Oncology (C.M.H., A.P.) and Pathology (L.R.), IWK Children's Hospital, Dalhousie University, Halifax, Nova Scotia, Canada.

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Address reprint requests to Dr. Allan Pyesmany, IWK Children's Hospital, 5080 University Avenue, Halifax, Nova Scotia, B3J 3G9, Canada.

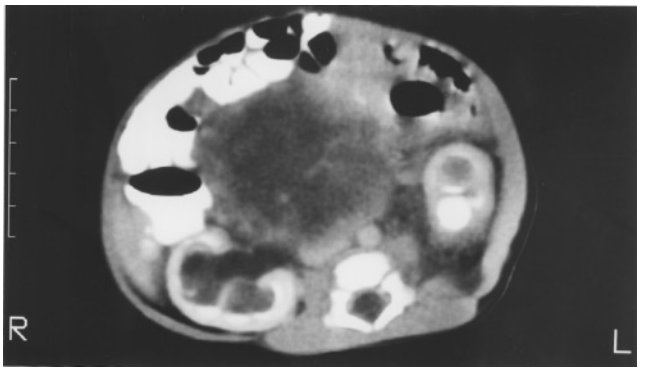

Fig. 1. Computed tomography of abdomen and pelvis shows a 6 by 10 centimetre retroperitoneal mass distinct from adjacent structures. No calcification is seen.

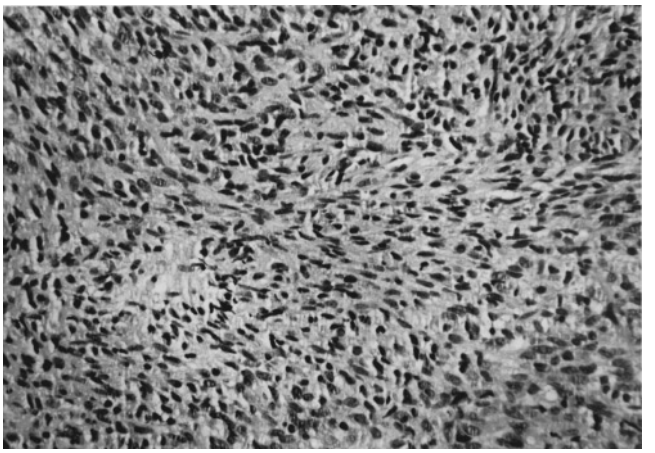

Fig. 2. Photomicrograph of original biopsy showing spindle cells arrayed in irregular ill-defined fascicles. The cells show only minimal pleomorphism. Hematoxylin and eosin stain. Magnification 200×.

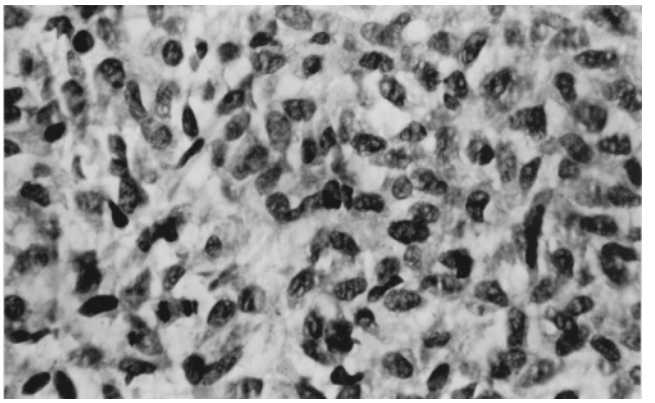

Fig. 3. Photomicrograph of original biopsy. The nuclei are round to oval with a vesiculated nucleoplasm and inconspicuous nucleoli. The cytoplasmic boundries are not discernible. A mitotic figure is present in the lower right quadrant. Hematoxylin and eosin stain. Magnification 500×.

diac dysrhythmia. On several occasions surgery was interrupted to permit external cardiac massage and cardioversion. Ultimately, 80% of the tumor was resected; deep attachments within the pelvis remained. At the time of surgery it was felt that the tumor's origin was retroperitoneal, although, because of its size, this could not be proven. A nephrostomy tube was inserted into the right renal pelvis prophylactically for obstructive uropathy.

Specimens of the tumor were obtained for gross and microscopic pathologic examination, light and electron microscopy, as well as immunohistochemical profile. Histopathological examination revealed a cellular tumor composed of ramifying vascular spaces surrounded by closely packed, spindle-shaped cells. In many areas the tumor cells showed no organization but in some regions there were aborted attempts at fascicle formation (Fig. 2). Greater than 10 mitotic figures were seen per 10 high power fields (Fig. 3). Tumor necrosis was seen on many sections. Immunohistochemical staining for vimentin was strongly positive. The electron microscopic examination appearances are similar to those previously described in fibrosarcoma [1]. Combination of the light microscopic, immunohistochemic and electron microscopic, findings are characteristic of fibrosarcoma.

Initial therapy consisting of 100% vincristine (0.05 mg/kg) and actinomycin D (0.0075 mg/kg/day) and 50% cyclophosphamide (37 mg/kg; VAC) was given 3 weeks after admission, 2 weeks postoperatively. This regimen was well-tolerated with virtually no myelosuppression. The nadir of white blood cells (WBC) was >6.0; platelets remained high at >700 and Hgb remained >11.0. It was decided to use 75% actinomycin-D and cyclophosphamide on the second course; however, the patient unfortunately did not respond well. He was given vincristine (0.05 mg/kg), actinomycin-D (0.0125 mg/kg) and cyclophosphamide (55 mg/kg). He developed life-threatening hepatic, hematologic and gastrointestinal actinomycin-D toxicity, but recovered within 10 days and was discharged in stable condition. A gallium scan subsequently showed continued uptake in the area of the tumor, of similar size as pretreatment, but a decrease in the density of uptake.

Week 6 of the chemotherapy was decreased to 50% due to toxicity. The following two cycles were well-tolerated. He was given slightly increased doses of 75% vincristine, 60% actinomycin-D and 60% cyclophosphamide. After 8 months of follow-up, our patient appears disease-free and is developing normally.

DISCUSSION

The diagnosis of abdominal fibrosarcoma is uncommon in infants and children. Of the 177 cases of congenital fibrosarcoma that we reviewed in the literature,

only 12 (7%) have been either abdominal or retroperitoneal. This malignancy occurs more commonly in males (60%) in whom it chiefly affects the distal portion of the lower and upper extremities (72%) [1]. Infantile, or congenital fibrosarcoma is generally considered to hold a favorable clinical course when compared to the adult form.

Chung and Enzinger [1] found in their review of 53 cases with infantile fibrosarcoma, that, of the five patients treated with only local excision or biopsy, all had died of their disease. Of those treated with radical or wide local excision with or without adjuvant therapy, all were alive at follow-up which averaged 6 years. One of nine children treated with amputation alone died. Two of the 53 patients had retroperitoneal tumors and one of these was treated with only radiation and died [1].

In 1977, Soule and Pritchard [2] reviewed 110 cases of infantile fibrosarcoma. Eight of the 110 patients had fibrosarcoma located in the trunk. It was not specified how many of these cases were retroperitoneal. In their series, none of the children had been treated with chemotherapy, but rather by local excision in the majority of cases. A small number had amputation and radiation and one patient had no treatment. The recurrence rate after local excision was 47%, and 13.5% developed metastatic disease. Treatment in 10 patients who died included primary excision alone (6 of 10), primary amputation (2 of 10) and primary radiation (1 of 10). Because Soule and Pritchard also divided their patients by age, they concluded that patients developing fibrosarcoma before the first 5 years of life have a 7.3% chance of dying of their tumor or of developing metastatic spread.

Studies prior to 1980 indicated that the most effective treatment of fibrosarcoma was total excision. However, more recent case reports are suggesting other forms of treatment are equally effective in some cases. Delepine [5] reported a complete response to chemotherapy alone using ifosfamide, vincristine and actinomycin D. There was no clinical evidence of a recurrence after nine months [10].

As late as 1990, Salloum et al. [9] reported 8 cases of congenital fibrosarcoma. Three patients were treated by local excision alone, one by surgery with radiotherapy, one by surgery with chemotherapy, one by radiation alone and two by all three modes of treatment. Of the two patients with retroperitoneal tumors, one was treated by surgery with chemotherapy and the other by surgery with radiotherapy. Both were alive and free of disease after 13 and 27 years of follow-up, respectively. Salloum et al. [9] noted that local recurrence seemed to have no relationship to the development of metastases. They also noted that in their two cases with distal recurrences, both had histological transformation to malignant fibrous histiocytoma (MFH). They recommended adjuvant chemotherapy to prevent distant metastases and the development of MFH. We suggest, however, that they do not substantiate their statement, "There is no evidence that adjuvant chemotherapy . . . improves local control," as all three patients in their study who received initial adjuvant chemotherapy were alive with no evidence of disease on follow-up. In addition to our own success with adjuvant chemotherapy, many other authors report success with chemotherapy. In response to Salloum et al.'s proposition, Brock et al. [7] reported an excellent response to four cycles of preoperative VAC in one of their patients.

Ninane et al. [3] described two newborns with huge unresectable tumours who responded so well to preoperative VAC every three weeks that complete surgical excision was possible after nine and six cycles of VAC, respectively. A third infant received postoperative VAC for residual microscopic disease. No recurrence had been noted after 6 years of follow-up in any of the three cases [3,6].

Robinson and Crawford [4] reported the successful use of intralesional excision (preoperative diagnosis was abscess) and a 21-month course of the VAC protocol in a massive congenital fibrosarcoma of the thigh in which excision was not possible due to vascularization and location. After 5 years of follow-up, the child was still cancer-free.

For completeness, we note that Spicer reports spontaneous regression of a fibrosarcoma of the forearm [10].

CONCLUSION

Chemotherapy appears to play an increasingly effective role in the treatment of congenital fibrosarcoma today, despite previous opinion that total surgical excision was the only effective treatment. After percutaneous biopsy of unresectable lesions, preoperative chemotherapy to reduce the size of the tumor may help to avoid mutilating surgery, and postoperative chemotherapy may permit complete resection after several months. Curative use of chemotherapy alone has been reported. Our case demonstrates that where excisional surgery is not possible, chemotherapy offers effective additional treatment.

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